Ruth Adewuya, MD (host):

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Ruth Adewuya, MD (host):

I am your host, Dr. Ruth Adewuya. This episode is part of our Hot Topics Miniseries. In this episode, we are covering trending clinical topics that have been in the news recently: monkeypox and Ramsay Hunt. First, we'll start with a conversation with Dr. Stan Deresinski on monkeypox. In the second part of the episode, Ishita Verma, a Stanford undergraduate student, an intern for Stanford Medcast, will be chatting with Dr. Jon-Paul Pepper on Ramsey Hunt syndrome.

Ruth Adewuya, MD (host):

Dr. Deresinski received his medical degree from the University of Illinois College of Medicine and received training in internal medicine there and at Stanford where he also completed a fellowship in infectious diseases. For three decades, he maintained a private practice in infectious disease, HIV, and travel medicine, and was a hospital epidemiologist at Sequoia Hospital where he also served as president of the medical staff for two years.

Ruth Adewuya, MD (host):

Dr. Deresinski is currently a clinical professor of medicine in the Division of Infectious Disease and Geographic Medicine at Stanford. He is the Medical Director of the Stanford Antimicrobial Stewardship Program and Chair of the Pharmacy and Therapeutics Committee and of the Specialty Drugs Subcommittee. He is a section editor of Clinical Infectious Diseases and is a past Chair of the Infectious Diseases Society of America, Standards and Practice Guidelines Committee as well as a member of the IDSA board of directors. Thank you for chatting with me today.

Stanley Deresinski, MD (guest speaker):

Well, thank you for having me.

Ruth Adewuya, MD (host):

Today we're talking about monkeypox. With what seemed like a sudden spread, monkeypox has felt like it has caught us off-guard and caused a little bit of panic, especially coming off the frenzy of COVID-19. Can you give our listeners a broad overview of monkeypox and its origins?

Stanley Deresinski, MD (guest speaker):

Sure. Monkeypox virus is one of more than 10 members of the genus viruses called Orthopoxviruses, which includes closely-related pox viruses, including, of course, smallpox. I might add, despite the name, it does not include chickenpox. Chickenpox is a herpes virus that has nothing to do with these pox viruses. The illnesses caused by these viruses range from lethal, usually lethal with smallpox, which, fortunately, doesn't exist in the wild anymore, to mild illnesses, for instance, with cowpox and the like.

Stanley Deresinski, MD (guest speaker):

The monkeypox virus is interesting. Even its name is interesting, in a matter of dispute, because it was first discovered in laboratory monkeys, macaques, in Denmark in 1958, two of them that had been imported from elsewhere. The virus was discovered then. That's how it got its name, monkeypox. As we now know, monkeys are not the prime reservoir of this virus. In fact, it isn't absolutely certain what is in the wild in Africa, but it seems to be small mammals, particularly rodents. Monkeys got a bad rap. They were first.

Ruth Adewuya, MD (host):

To what extent do you think the name monkeypox has fueled public interest in this virus?

Stanley Deresinski, MD (guest speaker):

In fact, WHO is discussing changing the name of the virus, perhaps for that reason and also because in relationship to the two versions of the monkeypox virus, because one version comes from West Africa and the other from Central Africa, and they have those names attached to them. There is a big movement to stop naming the viruses for geographic reasons because it maybe have an associated stigma, so those names in particular are likely to change.

Stanley Deresinski, MD (guest speaker):

I might also add that there's a distinct difference between the two clays of virus from those two sites in that the West African strain in Africa has a mortality rate of approximately 1%, whereas the Central African strain has one of about 10%. Fortunately, the strain we're seeing circulating throughout the world now, particularly Europe and North America, is the West African, less lethal strain.

Ruth Adewuya, MD (host):

You have mentioned that monkeypox is from the same family as smallpox and cowpox. I'm curious about the differentiation between those diseases and how monkeypox infectiousness compares.

Stanley Deresinski, MD (guest speaker):

Well, smallpox kills people. If you get further extreme, things like... or, for cowpox, produce, usually, just a single skin lesion at the point of inoculation with no spread. Monkeypox is in-between.

Stanley Deresinski, MD (guest speaker):

Fortunately, it's lethality in the current out-of-Africa outbreak has been minimal. In fact, the WHO has just stated that there's only been one death associated with it out of the 3,500 or so cases that have been identified. In fact, in California, where we've had something like 60 cases, as of two days ago, it was stated by the California Department of Public Health that none of them even required hospitalization.

Ruth Adewuya, MD (host):

What I'm hearing from you is that monkeypox is not quite as lethal. However, there are significant implications to somebody getting ill with monkeypox. For someone who's contracted monkeypox, what are some of the signs that they will have? What are some of the signs that should trigger them to go to a hospital?

Stanley Deresinski, MD (guest speaker):

Although there's been no great variability in the cases that have been seen in the United States, there may be an initial several-day-long prodrome with flu-like symptoms, basically, meaning fever, malaise, aches, and pains followed by the appearance of the rash, if you will. The rash had been typically said to appear first in the mouth and face, but in the current cases, we're seeing it all over, possibly because we believe a lot of the transmission occurs during body-to-body contact during sexual activity and the like.

Stanley Deresinski, MD (guest speaker):

Many cases are presenting with anogenital lesions as a consequence. The rash in this case can be scattered. The lesions tend to be of the same stage. That was used to be taken as a distinction between smallpox and, say, chickenpox because in chickenpox, you have all different stages of rash present at the same time, whereas in smallpox, they all seem to coordinate in terms of their stage of progression.

Stanley Deresinski, MD (guest speaker):

The lesions in chickenpox which, again, I will say has nothing to do with monkeypox, as like with herpes simplex, you see blisters with fluid. If, for instance, you wanted to get a sample, what we recommend is that the blister be unroofed to have this base be swabbed. Of interest is that the lesions in monkeypox are much deeper, and you can't unroof them. I mean, I suppose you could with a blade, but just by using a swab, you can't unroof them. When we get a sample, actually, for testing, we just vigorously rub the surface of the lesion.

Stanley Deresinski, MD (guest speaker):

Now there are others. Some may erode and have other manifestations as well. I might add that I mentioned a typical prodrome, but some patients don't have a prodrome. They first appear with a skin eruption. Many of them will subsequently develop some low-grade fever. They may have, depending on the site of infection... For instance, we see cases with perianal lesions and proctitis. Obviously, those would have their own symptoms.

Ruth Adewuya, MD (host):

Thank you. You just set up my next question for you, which is, what are the current methods of treatment that are available for monkeypox?

Stanley Deresinski, MD (guest speaker):

The preferred method is a drug called Tecovirimat, which is a pill that is recommended you take for 14 days. There's very limited experience with it, almost none in monkeypox, but it looks to be very effective in the test tube and from some other experiences. That's the preferred treatment.

Stanley Deresinski, MD (guest speaker):

You can't just write a prescription for it and send the patient to the local pharmacy. It's only available through the CDC on their own investigational, new-drug-use format. You have to contact them. It's basically recommended for treatment for patients... are at risk of having more severe disease.

Stanley Deresinski, MD (guest speaker):

I mean, at an extreme, if we have a severely immunocompromised patient who gets this would be one example or a patient who has critical site infection, such as having inoculated their eyes. That might be

another. It's certainly something to be considered, the safety data, which is limited but looks safe. It's a reasonable thing to get.

Stanley Deresinski, MD (guest speaker):

There are other drugs which have some activity, one of which is readily available, but it's intravenous, is called cidofovir. Its efficacy, I think, is probably going to be less than the Tecovirimat. There's an oral prodrug form of cidofovir, brincidofovir, which is still investigational. Also, another thing's been used... I mentioned that sometimes people can inoculate their eye. There are antiviral eye drops that are used for other things like herpes infection. There are, clearly, treatments available.

Ruth Adewuya, MD (host):

Can you talk about the smallpox vaccine and whether it is also effective against monkeypox?

Stanley Deresinski, MD (guest speaker):

What we know is that the childhood vaccination with smallpox vaccine appeared to produce lifelong immunity against smallpox. It's been quoted that it produces about 85% immunity against monkeypox. We know from certain experiences that although, for instance, the 2003 outbreak, which we may want to talk about, in the United States, that individuals who had been vaccinated could still get infected, but they had milder disease with many fewer skin lesions.

Stanley Deresinski, MD (guest speaker):

The smallpox vaccination appears to be effective, but, of course, we're not using that. There is a version of that vaccine, ACAM2000, which is available, but there's a preferred vaccine called JYNNEOS, which is available but much shorter supply. The reason why it is preferable, particularly if one were to go to a mass vaccination circumstance, which I don't think anybody is considering for this disease because the entire population is not at risk, is that the ACAM2000 contains a weakened form of the prior smallpox virus which is capable of replication.

Stanley Deresinski, MD (guest speaker):

It can cause complications related to the viral continuing to replicate after you inoculate someone in it. The JYNNEOS vaccine is a genetically-modified version. It's a live virus, but it can't replicate. It's believed those complications, which are the most severe, related to smallpox and ACAM2000 vaccination would not occur with JYNNEOS. It's the preferred vaccine, but it's in relatively short supply right now. What's being considered with it is ring vaccination, vaccinating people who have been known to be exposed, people who are named as contacts of a case rather than people in general.

Ruth Adewuya, MD (host):

It sounds to me that we are in a better place with monkeypox than we were with COVID-19 in terms of having some treatment options. Would you agree?

Stanley Deresinski, MD (guest speaker):

COVID-19 was a new virus. About every six months, it turns into another new virus, basically, which complicates things further. We had no diagnostics, no therapeutics, no nothing. In this case, this virus has been known, as I said, since 1958. It's probably been less studied than it should have been. I mean,

all of this is another example of how with transmissible infections, that their endemicity anywhere in the world is a potential danger to everyone and needs to be dealt with.

Stanley Deresinski, MD (guest speaker):

As I just mentioned, we have therapeutics. We have vaccines, and we know how to control it, which is basically behavioral modification. It generally requires close contact, often skin-to-skin contact for transmission, in contrast to SARS-CoV-2, the cause of COVID-19, which can be transmitted by aerosol at long distance with the viral particles persisting in the air for hours. It's a big difference. This is an eminently controllable disease.

Ruth Adewuya, MD (host):

You alluded to this earlier in our conversation, but I want to call it out specifically here. We know that monkeypox has been around for several decades, but there seems to be an upward spread right now, especially outside of Africa. Do you have any theories around what is causing that upward spread right now?

Stanley Deresinski, MD (guest speaker):

Over the decades, there's been an increased number of cases in Africa. It's believed that's largely related to two major factors. One is the encroachment of people into the areas where the putative animal reservoirs exist, and the other is the discontinuation of smallpox vaccination in the past because smallpox vaccination protected, as I said, at a rate of about 85% against monkeypox. Nobody who's young in Africa has been vaccinated. It's been increasing over the last years in Africa.

Stanley Deresinski, MD (guest speaker):

There have been, of course, cases over the years that have been transmitted outside of Africa by travelers and by other mechanisms. In 2003, there was an outbreak in the Midwestern United States that involved 47 people. The way that happened was that there had been imported Gambian pouched rats from Ghana to a pet distribution center where those animals came in contact with other animals and, particularly, prairie dogs. Who knew that people wanted pouched rats or prairie dogs for pets?

Stanley Deresinski, MD (guest speaker):

It was spread by the prairie dogs, which were sold to people throughout the Midwestern United States. As I said, 47 human beings were infected, either working at the pet distribution centers and pet shops or who had purchased the prairie dogs. It happens.

Stanley Deresinski, MD (guest speaker):

That outbreak is interesting from a number of points of view. Again, nobody died. In that instance, there were no secondary cases, which is very different from what we see now. I might mention, in 2001, there were two introductions in the individual travelers introduced to the United States. One of them, I think, also is very instructive.

Stanley Deresinski, MD (guest speaker):

This is a patient who flew from West Africa to Dallas, Texas, with a stop in Atlanta. A few days after arriving in Dallas, he presented to an emergency department with skin lesions. The remarkably astute

emergency room doctor figured out there was something strange here, contacted the CDC, and a rapid diagnosis of monkeypox infection was made.

Stanley Deresinski, MD (guest speaker):

The CDC, of course, did an investigation, and they identified something over 200 potential contacts. There were no high-level contacts, but a number of them were, for instance, people sitting on the airplane next to this guy and others, so people who drove him to the hospital. None of those people got monkeypox.

Stanley Deresinski, MD (guest speaker):

Again, the difference between that and what we're seeing now is skin-to-skin contact, I believe, in the current outbreak. There have been theories such as, could the virus been circulating at low levels, even in non-endemic countries, in animals and not been recognized? I suppose that's possible.

Stanley Deresinski, MD (guest speaker):

There were some very specific things that occurred early on that I don't know that they've been confirmed. There were two gatherings they've variously described as raves or whatever, one in Belgium and one in the Canary Islands. It's believed that somebody brought, probably to each of these sites, monkeypox. People there were infected, and then they dispersed to their homes. Further cases occur.

Stanley Deresinski, MD (guest speaker):

Basically, if you will, perhaps it's probably a stretch to call it that, but superspreader events with subsequent dissemination. Then a dissemination occurs through what have been described in investigations of sexually-transmitted disease as strong sexual networks with the obvious means of spread.

Ruth Adewuya, MD (host):

Thank you for that insightful response. As we wrap up our conversation, I'd like to end by asking you, what advice would you have for clinicians who are getting questions from their own patients about monkeypox? What are some of the next steps that they can take?

Stanley Deresinski, MD (guest speaker):

Well, the CDC has excellent information at their site freely available, and they're constantly updating it. Right now, that's the best source of information. They can also contact their local public health department if there are particular issues and the like. In terms of just getting the general information and instructions, the CDC websites are great.

Ruth Adewuya, MD (host):

Fantastic. Thank you so much for jumping on and sharing your insights on this topic. I'm sure that as things evolve, I would like to continue this conversation if there's new information to share. Thank you for-

Stanley Deresinski, MD (guest speaker):

All right.

Ruth Adewuya, MD (host):

Taking the time to chat with me.

Stanley Deresinski, MD (guest speaker):

Thank you.

Ishita Verna (host):

Dr. Pepper is a double-board certified surgeon who specializes in aesthetic and reconstructed surgery of the face, in particular surgery for the treatment of facial paralysis. He is the doctor of the Stanford Facial Nerve Center since 2017. He has board expertise in facial plastic and reconstructive surgery, including facial reanimation surgery, facelift surgery, rhinoplasty, and the reconstruction of the face after skin cancer resection.

Ishita Verna (host):

Dr. Pepper performed his undergraduate studies at Brown University, majoring in neuroscience. He was awarded his MD at the University of California, Irvine, graduating with the highest honors and Alpha Omega Alpha designation in 2007, and went on to the University of Michigan for his residency training. He now directs the scientific work of the Stanford Facial Nerve Center. His NIH-funded research explores regenerative strategies to improve nerve regeneration after injury.

Ishita Verna (host):

Thank you so much for chatting with us today, Dr. Pepper. As you know, Ramsay Hunt syndrome, or RHS for short, has very much been a trending topic recently. However, since it is a relatively rare condition, not many people know in detail about this type of facial paralysis. Dr. Pepper, to get us started, do you mind describing the condition and its symptoms?

Jon-Paul Pepper, MD (guest speaker):

Yeah. First of all, thank you so much, Ishita, for having me on as a guest today. Thank you, Dr. Adewuya, as well. Yeah, Ramsay Hunt syndrome... Obviously, a little bit more on people's minds in light of recent events. In summary, Ramsay Hunt syndrome describes a relatively rare presentation of a disease where a patient has facial paralysis, usually pain in some part of the face, particularly the ear, and then importantly, a rash that looks like a chickenpox rash that breaks out on some surface of the head and neck, most commonly, the ear, sometimes inside of the mouth.

Ishita Verna (host):

What might be some of the causes for this condition?

Jon-Paul Pepper, MD (guest speaker):

Ramsay Hunt syndrome is caused by the varicella virus, the same one that causes chickenpox. This is a story... As many things in medicine are, it's all about location, location, location. Most of us are exposed to herpes varicella or the chickenpox virus at some point in our life, but many viruses are neurotropic. They like to either go after and live in nerves or have some attraction for nerve tissue.

Jon-Paul Pepper, MD (guest speaker):

The herpes varicella virus is similar to this. It will hide out for long periods of time in the sensory nerves, most likely, the sensory nerves, and then will manifest itself later in life. Commonly, patients in their 50, 60, 70s, maybe even 80s, will have what we commonly call the shingles, right? That's where you get pain in some sensory nerve distribution somewhere on your body. You also get the chickenpox-like rash, oftentimes.

Jon-Paul Pepper, MD (guest speaker):

For Ramsay Hunt syndrome... Again, this is about location, right? It happens to be harbored in the distribution of the facial nerve or some of the adjacent nerve tissue. It manifests itself as pain. Particularly, ear pain or some type of hemifacial pain would be common, a chickenpox-like rash may break out, particularly, over kind of the conchal bowl of the ear, the complex surface of the ear that we process sound through, and then ipsilateral facial paresis. Those would be the hallmarks. That's the kind of who, what, why, and where.

Ishita Verna (host):

We talked about some of the functioning and physical difficulties that patients encounter. Can you also walk us through some of the social and psychological issues that patients might encounter?

Jon-Paul Pepper, MD (guest speaker):

Certainly. I think this gets maybe not as much attention because the initial presentation can be frightening, right? Many times, people may wonder if they're suffering a stroke because it's acute onset, and they lose function of one side of their face. That's very off-putting and very alarming. However, the recovery from this takes a long time, even when adequately treated.

Jon-Paul Pepper, MD (guest speaker):

Most full studies of Ramsay Hunt syndrome would last about a year to find the final result of recovery from facial paralysis. That's a long time to have your face not working normally. There's been, in the last 10 to 20 years, a growing and very important body of research, a small, small fraction of which I have done or with collaborators, so people that I've worked with, that's looked at the psychosocial and also communication aspects of facial paralysis. They are profound. It's a very impactful disease from that respect.

Ishita Verna (host):

With that in mind, what are the possible treatment options that are available for patients?

Jon-Paul Pepper, MD (guest speaker):

When a patient presents acutely, the standard form of treatment... First off, diagnosis may not be easy, depending on how they present. That may take some time, to make a diagnosis. Presuming that it's a classic triad with pain, facial paralysis, and a vesicular rash, the standard treatment would be steroids, usually oral steroids, and then critically high-dose antivirals.

Jon-Paul Pepper, MD (guest speaker):

This is an area of some controversy. There have been prior Cochrane Reviews that have had some difficulty demonstrating benefit for high-dose antivirals, but based on the best existing data, my

recommendation is that anti-inflammatories with high-dose PO steroids in combination with high-dose antivirals are the foundation of treating it acutely.

Ishita Verna (host):

It sounds like there's a lot of treatment options available for Ramsay Hunt syndrome. Do most patients tend to make a full recovery, or do we see any long-term healing outcomes that might still consist?

Jon-Paul Pepper, MD (guest speaker):

In general, the standard for comparison here is Bell's palsy, which is kind of the elephant in the room. I think for anyone who treats patients with acute facial paralysis, the things that are on their mind are, number one, stroke, making sure it is not a stroke. Number two, Bell's palsy, once stroke and other causes are eliminated as the potential cause of the patient's acute-onset facial paralysis.

Jon-Paul Pepper, MD (guest speaker):

Bell's palsy is very common. Then Ramsay Hunt is a subset of those patients with acute-onset facial paralysis. It's a much smaller component. To compare, how do these patients do... About 10% or so of Bell's palsy patients don't get a full recovery. That number depends a little bit on how closely you're looking at them. It might be a little bit higher, maybe 10 to 15% or so of patients don't recover to what maybe passive observers might deem normal.

Jon-Paul Pepper, MD (guest speaker):

Ramsay Hunt syndrome patients, though, tend to do worse, on average. We don't have near the number of them for a study, but they certainly appear to recover more poorly than the average Bell's palsy patient. That's not to say, though, that an individual can't make a full recovery, right? That just means on average. We take hundreds and then thousands of patients and then try to calculate averages, which is difficult to do.

Jon-Paul Pepper, MD (guest speaker):

In any event, it seems clear that the average recovery from Ramsay Hunt syndrome is worse than that of a patient with Bell's palsy. The reason for that is the intensity of the inflammation, most likely, and the degree of nerve injury. Ramsay Hunt syndrome, much more frequently than Bell's palsy, involves a second cranial nerve or multiple cranial nerves. That's a testament to a high degree of inflammation that spreads and belong multiple neural pathways.

Ishita Verna (host):

It seems like the distinction between Bell's palsy and RHS is really important. Could you talk us through some of the more nuanced similarities and differences of the two conditions?

Jon-Paul Pepper, MD (guest speaker):

Bell's palsy is a diagnosis of exclusion. It is technically idiopathic, acute-onset facial paralysis. It's also thought to have a viral etiology of viral cause, but they are exclusive terms. Ramsay Hunt syndrome is a manifestation of the chickenpox virus reactivating, also causing the other symptoms we discussed previously, but they can present somewhat similarly, aside from the rash and the hallmark of sometimes intense pain that's associated with Ramsay Hunt.

Jon-Paul Pepper, MD (guest speaker):

They are separate diagnoses, and they're treated, again, similarly but a little bit different. This is an important distinction. Bell's palsy patients are usually treated following accurate diagnosis and whatever workup is needed. Bell's palsy patients are usually treated with high-dose steroids frequently, say, a 10-day course or something like that is what's commonly used and is supported in the literature.

Jon-Paul Pepper, MD (guest speaker):

Also, I include a prescription for an antiviral. I will usually use Valacyclovir to treat Bell's palsy as well. There are some randomized control trials that show some incremental benefit when one includes lower-dose Valacyclovir along with steroid treatment.

Jon-Paul Pepper, MD (guest speaker):

Let's then shift to Ramsay Hunt syndrome. Okay, there's a difference here between, again, the intensity of the inflammation. Steroids do appear to be beneficial for the treatment of Ramsay Hunt syndrome. There's a little bit of controversy around this, but, in general, for people who treat facial palsy, steroids would be included in the treatment of Ramsay Hunt syndrome.

Jon-Paul Pepper, MD (guest speaker):

I feel very comfortable saying that, but the dose of antiviral is different. It has to be escalated quite a bit. Ramsay Hunt syndrome is treated with high-dose antivirals. This is where making an accurate diagnosis is really critical, that one has to accurately figure out, this is actually Ramsay Hunt syndrome, and high-dose antivirals may be needed.

Jon-Paul Pepper, MD (guest speaker):

Also, the patients can be in intense pain. They may be very uncomfortable. Initial period of in-patient observation is sometimes considered. Especially if they present acutely, they may be in excruciating pain. That may require pain medicine, for instance, to just help them be comfortable and get sleep. They also are going to need high-dose antivirals. They need to be able to take medicines by mouth. If not, it might have to even be given intravenously. They are treated very differently, and it's important to make that distinction.

Ishita Verna (host):

There are several key fundamental differences between the two conditions. When a clinician is trying to diagnose and differentiate between the two, what are some key indicators that they should look for for Bell's palsy and for RHS?

Jon-Paul Pepper, MD (guest speaker):

The rash is a big key because it's right in front of you. A patient presents with acute-onset facial paralysis. They will tell a story. Let's do scenario one. The patient with Bell's palsy, maybe who comes into urgent care, emergency department, or their primary care physician for evaluation... They should, if this is Bell's palsy, tell a story of acute onset, like, "Doc, I woke up with this, and then the left side of my face wouldn't move."

Jon-Paul Pepper, MD (guest speaker):

Importantly, the brow is also paralyzed. We're taught this from a young age in medical school, that central nervous system etiology, such as stroke, may preserve brow function. The location of the nerve injury does matter here, or the nerve tissue injury does matter. For Bell's palsy, the brow is also affected. There is paralysis of the ipsilateral frontalis muscle causing brow paralysis, as well as the rest of that ipsilateral side of the face. It's acute onset.

Jon-Paul Pepper, MD (guest speaker):

There is no rash associated with it. There may be pain, but that's plus, minus. The Ramsay Hunt syndrome patient may tell you a different story. They may say, "I had the pain first." They may tell a story of pain that precedes the other manifestations of it.

Jon-Paul Pepper, MD (guest speaker):

Importantly, though, the rash. If the rash is present in the conchal bowl, or on the auricular surfaces, or the ear surfaces, that's a very helpful diagnostic point. One must remember to check inside the mouth. The oral mucosa may be... I have seen patients such as this, where they have ipsilateral facial paresis, hemifacial pain, or periarticular pain, but then the only location of the rash is inside the mouth, particularly, the soft palette, because that receives some sensory innervation from the facial nerve more proximally. The rash is key, but one must remember to check inside the mouth for that rash.

Jon-Paul Pepper, MD (guest speaker):

There's one other important nuance, though, is that there's a phenomenon of Ramsay Hunt syndrome appearing without the rash, so herpes zoster sine herpete. That's a difficult diagnosis to make. The patient has unilateral facial paralysis but no rash, even when checking inside the mouth. How can one distinguish this diagnosis?

Jon-Paul Pepper, MD (guest speaker):

There's no definitive way to do it, but your index of suspicion must be high when the patient has intense pain. The patient who has intense, boring otalgia, right... They have, again, acute onset of their facial paralysis. There's no rash makeup, but their pain is intense. In those patients, I have a higher index of suspicion that they may have herpes zoster sine herpete in the facial nerve. That, though, is a very challenging diagnosis. Really, the key for all of us, kind of the average circumstance facing a clinician, key would be the rash and facial palsy.

Jon-Paul Pepper, MD (guest speaker):

I did mention briefly, earlier that Ramsay Hunt syndrome is known to cause, oftentimes, second cranial neuropathies or additional nerves that are injured. That would also increase your suspicion. Although that can happen with Bell's palsy, it would increase your suspicion that maybe you're dealing with Ramsay Hunt syndrome.

Jon-Paul Pepper, MD (guest speaker):

Hearing loss can be associated with Ramsay Hunt syndrome. There can be other cranial nerves affected with Ramsay Hunt syndrome at a slightly higher frequency than Bell's palsy. Those would also maybe be cues to make you think a bit more about Ramsay Hunt syndrome, if they were presenting very atypically, maybe with multiple cranial nerves.

Jon-Paul Pepper, MD (guest speaker):

Lastly, whenever there's more than one cranial nerve affected, whether it be the facial nerve and a suspected diagnosis of Bell's palsy, but also hearing loss, or if the suspicion is maybe this is Ramsay Hunt, but you're not clear on there being a rash, and there's tongue weakness plus the facial paralysis, these patients merit imaging. They're best served by an MRI with contrast because in those cases, we're also trying to rule out a central nervous system neoplasm.

Jon-Paul Pepper, MD (guest speaker):

The clinical practice guidelines that we use that are very helpful and succinct for Bell's palsy dictate that if you have two cranial nerves affected, you need to obtain imaging, contrasted imaging, I would add. You need intravenous contrast to show a tumor. A non-contrasted study is less helpful if that's what you're looking for. Patients with Ramsay Hunt, particularly if you can't see a rash, they need to be imaged.

Ishita Verna (host):

As we're finishing up, do you have any last insights to share with our audience?

Jon-Paul Pepper, MD (guest speaker):

I think that Ramsay Hunt syndrome, obviously, is getting a little bit of its time in the spotlight just due to recent events. I think, though, that it points at several things. One, things that present with acute neurological change are frightening. They're frightening for patients, but let's be honest. They're unsettling for us as clinicians as well.

Jon-Paul Pepper, MD (guest speaker):

I think, number one, having a very strong foundation... If you're someone who faces acute problems, having a strong foundation in Bell's palsy is really the core. Knowing exactly what Bell's palsy is really helps with the when, and the why, and the how you manage other more low-frequency, facial palsy presentations.

Jon-Paul Pepper, MD (guest speaker):

I would say that's always the starting point. Bell's palsy is sort of the foundation. Then that is not Ramsay Hunt. That's an important point to remake and revisit, is that Bell's palsy is not the same thing as Ramsay Hunt syndrome. Then we talked in detail about the diagnostic criteria, the way patients would present with Ramsay Hunt and then their recovery.

Jon-Paul Pepper, MD (guest speaker):

I think the final point is people may want to know with, say, a Ramsay Hunt patient. Patients should not be trying to be seen by a subspecialist acutely. They need to be seen quickly. They usually do because they're very concerned that half of their face no longer moves. They think they're having a stroke, but you may have the odd patient who knows someone with Bell's palsy and somehow minimizes it. It may be more up to the provider to act quickly. The patients need quick medical treatment.

Jon-Paul Pepper, MD (guest speaker):

Going back to the analogy of a finger getting crushed with a titanium ring around it, time is of the essence, right? As the swelling progresses, the tissue injury can increase. Those patients need to be seen

rapidly by a local emergency department, in their PCP's office, or in urgent care, whatever they can get access to quickly. That's the key for all of these presentations.

Jon-Paul Pepper, MD (guest speaker):

When would they see someone like me? Sometimes in the first three months after injury, I may enter into caring for these patients to help them plan out, how do we forecast recovery, what time intervals are important, but I wouldn't be deciding on any surgical treatment for, usually, roughly a year.

Jon-Paul Pepper, MD (guest speaker):

In my clinic, I will see them first at three to six months. I'll connect them with physical therapists, people who can help them with their recovery. If they need electrical testing of their nerve, I refer them to specialized neurologists who perform that service. That process can begin in the three-to-six-month time window, but it's not the same for every person.

Jon-Paul Pepper, MD (guest speaker):

Actual treatment from me usually begins at about one year after treatment because I want to see how much they get better on their own. It is very hard even with modern surgery to beat, to do better than native recovery, right? That nerve is going to try to recover. Let's see what it can do before we start supplementing it with surgery. By and large, I won't intervene surgically for at least one year.

Jon-Paul Pepper, MD (guest speaker):

In my opinion, there's not necessarily much harm in waiting a little longer, if needed. If the trajectory of healing looks a little promising, it's not like we have to make a decision right at one year for a patient with Ramsay Hunt.

Ishita Verna (host):

Thank you so much for all the information. I know I definitely learned a lot, and I hope our audience did as well. Again, thank you so much.

Jon-Paul Pepper, MD (guest speaker):

It's my pleasure.

Ruth Adewuya, MD (host):

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